Portal HTN

DEF	↑↑ portal venous pressure>12 mmHg.		
AE	Pre-hepatic	- Umblical:sepsis, UVC. - Portal vein: Thrombosis,,,agenesis, atresia,,AV Fistula. - Splenic vein: Thrombosis - Trauma, sepsis,	
	Intra-hepatic	Pre-sinusoidal (portal tract)	- Fibrosis(bilharziasis, congenital). - Infiltration(leuk, lymph.).
		Sinusoidal	- Cirrhosis.
		Post- sinusoidal	- Veno-occlusive diseases.(VOD)
	Post-hepatic	- Budd-Chairi S - IVC obstruction(thrombus/ mass). - CVS: Congestive HF, Constrictive pericarditis, pericardial effusion.	
Pathology& effects	Congestion	Viscera drained by portal vein(SM, ulcers, malabsorption)	
	Opening	Porto-systemic collaterals(esoph.+ anorectal varices, caput medosa,).	
	LCF	Edema, ascites, encephalopathy.	
	Ascites	↓↓Albumin, Na^+ H_2O retention(↑↑aldosterone, ADH), Lymphorrhea.	
C/P	- AE.,,,,,HSM.,,,,Ascites.,,,,,bleeding varices(haematemesis, melena). - Dilated veins+ C/P of LCF.		
Invest	Lab	LFT, CBC	
	Rad	Barium, U/S, CT, MRI, MRA, Transhepatic& spleno-portography,	
	Invasive	- Endoscopy, portal manometry(wedged hepatic venous pressure). - Liver biopsy.	
TTT	Portal HTN+ Active GIT bleeding	- ABC. - Monitor vital signs. - IV line - Anti-shock. - Vit K, PLT, Blood. - H2 Blockers. - NGT/ syngestaken tube+ Stomach wash. - TTT of encephalopathy. - Vasopressin/ Glypressin/Octreotide(splanchenic VC). - Endoscopic(sclerotherapy/ band ligation). - Surgical(porto-systemic shunt/ vasoligation).	
	Prophylactic	- Medical: Propranolol(1-4mg/kg/d). - Endoscopic:sclerotherapy. - Surgical:shunt - Liver transplantation.	

Approach to portal HTN

HX			NICU Admission(AE), Exchange transfusion, bleeding…..etc.
Exam	Splenomegaly		Most important sign of portal HTN.
	Liver	NO HM	Prehepatic AE.
		Shrunken	Cirrhosis.
		Maeked	Post-hepatic.
	Ascites	Cirrhosis, LCF.	
Invest	LFT(Normal in prehepatic& presinusoidal).		

Liver Cirrhosis

Def		Chronic, diffuse, irreversible liver disease(degeneration of liver cells& irreversible loss of liver architecture)
AE	Infection	Post hepatitis.
	Drugs	INH, MTX.
	Toxins	Alcohol
	CVS	Cardiac cirrhosis
	Biliary	Atresia
	Metabolic	Wilson, α1ATD, Haemochromatosis, GSD IV, Galactosemia, tyrosinaemia.
	Immune	Auto-immune hepatitis.
Classification		- AE. - Pathology(mico/macronodular,,,mixed) - Functional: compensated/ not.
C/P	Compensated	Accidentally discovered.
	Decompensated	- LCF,,,,Portal HTN,,,,HCC - Exam: firm,,,shrunken,,,sharp border.
Invest	AE	Hepatitis markers
	Lab	CBC, LFT.
	Imaging	U/S, CT, MRI.
	Invasive	Biopsy.
	For Portal HTN	As before.
	complications	αFP, U/S, CT, MRI.
TTT	- Ae,,,,LCF,,,, Portal HTN,,,,Complications(encephalopathy, Ascites, HCC) - Liver transplantation.	

Liver cell failure

Def	Impairement of liver function		
C/P	General		- AE. - Anorexia, fatigue,FTT. - Fever: low grade(bacteraemia).
	Mouth		- Foeter hepaticus.
	Skin,MM		- Jaundice. - Palmer erythema. - Spider nevi.
	Ascites& edema		- ↓↓Albumin,↑Na, H2O, Portal HTN,Lymphorrhea, peritonitis.
	Endocrinal	M	- Gynecomastia.
		F	- Amenorrhea.
	Encephalopathy		
	CVS		HDCirculation, porto-pulmonary shunts.
	Blood		Anaemia, bleeding, hypersplenism, pancytopenia, bleeding tendency.
	Renal		Hepato-renal S.
	Infection		Spontaneous bacterial peritonitis.
	Metabolic		Hypo/hyperglycaemia.

Hepato-Renal syndrome

DEF	Progressive functional RF in Ptn with LCF.
AE	Unclear(↑↑sympathetic→↑renal venous pressure→V.C→↓GFR→Oliguria& ↑creatinine.
Prognosis	90% mortality.
TTT	Liver transplantation.

Hepatic encephalopathy

Ae	↑↑	Neuro-toxins(GABA,,,Ammonia,,,Mercaptans): →→↓ cerebral O2 consumption→→coma. - ↑Protein diet,,,,old blood,,,,GIT bleeding,infection, surgery.			
	↓↓	K, alkalosis(vomiting, diarrhea, diuretics,,,,paracentesis)			
C/P(stages)		I	II	III	IV
	Symptoms	Lethargy	confusion	stuper	coma
	Signs	Drawing figures	Fetor hepaticus+ Asterixis	Hyper-reflexia + Asterixis	Areflexia (no Asterixis)
	EEG	Normal	Slowing	Marked abnormal	Marked abnormal/ silence

TTT	**General**	- Avoid PPT Factors - care of comatosed child.
	Diet	- ↓↓ protein. - ↑↑calories(CHO).
	Stomach	- Wash
	Lactulose	- Oral+ enema(↓↓ ammonia production& absorption+ osmotic laxative).
	Abs	- Neomycin(NGT).

Reys Syndrome

DEF	*Acute non inflammatory encephalopathy+ fatty degeneration of the liver documented by: - Clinical:↓consciousness. - Lab:CSF(<8 Leukocytes/mm^3). - Autopsy: brain edema without inflammation.
AE	Unknown (viral, aspirin, mitochondrial cytopathy).
C/P	- Age: 4-12 years old. - Prodromal URTI. - Later: Vomiting ±hypoglycemia.,Encephalopathy, Moderate hepatomegaly, no jaundice.
DD	- CNS infection. - Drug ingestion. - Hemorrhagic shock with encephalopathy. - Metabolic disease e.g. fatty acid oxidation, organic academia, urea cycle defects.
Invest	- ↑↑: Ammonia, Liver enzymes, LDH, CK- - ↓↓: hypoprothrombinaemia, Hypoglycemia - Liver biopsy→→Fatty infiltration, specific mitochondrial morphology on E/M.
TTT	- No specific treatment. - Supportive treatment(positioning, MV, IVF, DEXA, Mannitol, Phenobarbitone, cooling body .

	α 1 AT↓↓	Wilson disease. (Hepato-lenticular degeneration)
AE	AD	AR(↓↓ ceruloplasmin→→IEM of Cu.
Pathology	Unclear(α 1 AT is aprotease inhibitor so if↓↓→→ sever inflammation + tissue damage.	- Defect in biliary Cu excretion (accumulate in liver, brain, cornea, kidney).
C/P	- Liver: cholestasis+ HSM→→Cirrhosis+ portal HTN. - Lung(adult):emphysema(panacinar).	- Liver: HSM, hepatitis, cirrhosis, portal HTN, LCF. - Brain:tremors, sezuires, dysarthria, dystonia, migraine. - Blood: haemolytic anaemia. - Renal: fanconi like S/ RF. - Cornea: Kayser-Fleisher rings.
Invest	- Sr. α 1 AT→↓↓ - DNA study. - Biopsy:PAS +VE granules.	- ↑↑urine& Sr.Cu,,,,↓↓ ceruloplasmin. - Penicillamine chalange test. - Biopsy:LCF, Cirrhosis+ Cu accumulate in liver, brain, cornea, kidney. - Slit lamp. - CT, MRI.
Complications	- Cirrhosis, LCF, portal HTN.	
TTT	- Complications. - Liver transplantation.	- Diet:↓↓Cu. - Chelators: Penicillamine(nephrotoxic, BM toxic). - ↓↓Cu absorption: Zinc sulphate. - VIT E(Antioxidant. - Liver transplant.

	Budd- Chiari S	Veno-occlusive disease.
DEF	Hepatic vein obstruction→→ portal HTN (post-sinusoidal).	- Toxins: alkaloids, teas(jamaikan type). - Chemotherapy. - Infections& malnutrition.
AE	- Idiopathic - Thrombosis: SLE, Infection, polycythemia.	
C/P	- Acute:HM, Ascites, LCF, death. - Chronic:HSM, portal HTN.	* 3 stages: - Acute:Rapid ascites+dilated veins+ HM(no SM). - Subacute: Mild ascites& mild HSM. - Chronic: ascites, dilated veins, shrunken liver, LCF, SM(++)
Invest	- Venography. - Measure portal venous pressure.	- Liver: LFT, Biopsy, U/S, portal HTN. - Ascites: taping(transudate+↑↑ protein+ no PMNL).
TTT	- LCF, portal HTN, Ascites, Cs.	

Indications of pediatric liver transplantation

Obstructive Bililary Tract Disease:	- Biliary atresia . - Sclerosing cholangitis . - Trumatic . - Post -surgical .
Metabolic Disorders	- α 1-Antitrypsin deficiency . - Tyrosinemia type 1. - GSD-IV. - Wilson disease. - Primary oxalosis . - Neonatal hemochromatosis . - Crigler -Najjar type 1. - Familial hypercholesterolemia. - Organic acidemia . - Urea cycle defects.
Intrahepatic Cholestasis	- Idiopathic Neonatal Hepatitis - Alagille syndrome
Acute Hepatitis	- Fulminant hepatic failure . - Viral . - Toxin . - Drug induced.
Chronic Hepatitis with cirrhosis	- Hepatitis B or C,,,Autoimmune.
Primary liver Tumers	- Benign tumers (hamartomas , hemangioendothelioma). - Unresectable hepatoblastoma. - HCC.
Miscellaneous	- Cryptogenic cirrhosis. - Congenital hepatic fibrosis. - Caroli disease. - Cystic fibrosis. - Cirrhosis indused by TPN. - Polycystic liver disease.

Liver biopsy

Indications	- Neonatal cholestasis. - Chronic hepatitis. - Metabolic: galactosemia. - Storage diseases:GSD, Wilson, haemochromatosis. - Reys S. - HM(unexplained).
Contra-indications	- Bleeding tendency. - Vascular/ infected lesions. - Sever Ascites.
Post care	- Position: Rt side. - Monitor: vital signs. - Bleeding: IVF, FFP, PLT, blood+ U/S.
Complications	- Hge - AV Fistula. - Pneumothorax. - Infection. - Bleeding.

Fulminant hepatitis

DEF	Development of signs of LCF(Hepatic encephalopathy) within 8 wks of the onset of liver disease, in the absence of previous liver disease.	
AE	**Rapid deterioration of liver functions due to: - Infections:CMV, EBV, hepatitis A B C, adenovirus, toxoplasmosis - Metabolic: galactosemia, tyrosinemia, wilson's & mitochondrial disease. - Ryes S - Toxins &medications: INH, anticonvulsants, Acetaminophen. - Autoimmune: hepatitis,SLE. - Others: Ischemia, Mg.	
C/P	**Acute**	- Jaundice, vomiting, abdominal pain, bleeding, ascitis, HSM,
	Complications	- Encephalopathy , Hge, sepsis.
Invest	**Lab**	- Hypoglycemia - Electrolyte& Acid-base disturbance:↓↓Na, K, Ca,…etc. - Coagulopathy (↑↑ PT, APTT, INR) - ↓↓→→Hypoalbuminemia & hypoprotinemia - ↑↑→→ammonia, liver enzymes, bilirubin - KFT: RF
	EEG	- Encephalopathy(II-IV).
	CT	- Brain edema.
TTT		- Assess ABCs and admit to ICU &Consult gastroenterologist on call - Asses neurological status and level of consciousness (GCS≤ 7 →MV) - **Fluids**: 2/3 maintenance, 10-20% **dextrose** according to blood glucose. - Correct electrolyte (avoid hypertonic saline in hyponatremia, which can worsen hepatic encephalopathy) - **Vitamin K1**: Infants 1-2 mg/dose IV, Children 5-10 mg/dose IV - **Lactulose** (aim for 2 – 3 stools/day): Infants 2.5 ml /12h PO Children 5-10 ml /8h PO - **Ranitidine** 2 – 4 mg/kg/day IV /12h - IV antibiotics if indicated (not prophylactic) - **FFP**: <u>for DIC or active bleeding;</u> to avoid masking worsening liver function by correcting coagulation parameters. - **Mannitol** infusion if ICP is suspected - **N-acetylcystine** :for acetaminophen toxicity.
Follow up and monitoring:		- LFT, KFT, Electrolytes, bleedind& coagulation profile, glucose, ammonia, bilirubin/12-24h; depend on clinical situation. - CT: cerebral oedema - EEG: encephalopathy grading - Abdominal U/S

Upper GIT Bleeding

DEF	**Hematemesis**	Passage of vomited material that is coffee grounds in colour or contains frank blood.
	Melena	Passage of black tarry stool (bacterial degradation of hemoglobin).
AE	• Swallowed maternal blood (nipple fissure in breast feeding mother) • Esophagitis, gastritis, duodenitis and stress ulcers • Vascular malformation, aorto-esophageal fistula, esophageal varices • Coagulopathy, Vitamin K deficiency • Foreign body • Non GI causes (hemoptysis)	
Invest	• CBC, ESR • Coagulation profile • LCT.	
TTT	***ABC& support, Monitor + Pediatric gastroenterologist on call *****For stable patients:** NPO, start on IVF -IV ranitidine + Observe for 24 hrs *****For unstable patients: ICU+ monitor:** - Two IV lines should be placed - Urgent blood grouping and cross matching - IVF **0.9% NS 20ml/kg bolus** and can be repeated. - Saline lavage : asses bleeding, view source(esophageal varices: **- IV Ranitidine** 3-4 mg/kg/day q6-8h, maximum: 50 mg/dose **- Somatostatin or Octreotide** Dose: 1 µg/kg IV bolus then 1 – 5 µg/kg/h IV continuous infusion diluted in D5W or NS Indications: patient with upper GI bleeding 2ry to esophageal varices . Mechanism of action: it is a long acting synthetic analogue of Somatostatin (splanchnic VC). - After stabilization:**Upper GI endoscopy**. - If uncontrolled Hge : urgent endoscopy (varical injection or ligation).	

Clinical approach to Ascites:

Isolated	With general edema.
- Portal HTN. - TB, Mg, SBP.	- Renal:NS. - GIT: Protein loosing enteropathy. - Nutritional:PEM. - Liver: LCF

Ascites

<table>
<tr><td>DEF</td><td>Fluid accumulation in peritoneal cavity.</td></tr>
<tr><td>AE</td><td>- Portal HTN.
- ↓↓ Albumin: LCF, NS, Protein loosing enteropathy.
- Peritonitis
- Trs: lymphoma, neuroblastoma.
- Chylous: cong,anomalies,,,injury, surgery, lymphatic obstruction(mass).
- Biliary: perforation of CBD.
- Urinary: perforation of UT.</td></tr>
<tr><td>C/P</td><td>- AE.
- Exam
<table>
<tr><td>Inspection</td><td>Palpation</td></tr>
<tr><td rowspan="2">- Distension.
- Dilated veins.
- Divercation of recti.
- Skin:stretched, glistening,
- Umbilicus: everted, downward.
- Wide subcostal angle.</td><td>- Mild: dullness aroun umbilicus.
- Moderate:shifting dullness.
- Sever:transmitted thrill.</td></tr>
<tr><td>Auscultation:venous hum(Portal HTN).</td></tr>
</table></td></tr>
<tr><td>DD</td><td>- Fat, Faeces, Flatus, Fullbladder.</td></tr>
<tr><td>Invest</td><td>- AE.
- Tapping: physical, cells, chemical, bacteriological…etc.</td></tr>
<tr><td>Types</td><td>
<table>
<tr><td colspan="2"></td><td>Transudate</td><td>Exudate</td></tr>
<tr><td colspan="2">Aspect</td><td>Clear</td><td>Turbid</td></tr>
<tr><td colspan="2">Sp.gravity</td><td>↓↓(<1015)</td><td>↑↑</td></tr>
<tr><td colspan="2">Ptn</td><td>↓↓(<2.5gm)</td><td>↑↑</td></tr>
<tr><td rowspan="2">Cells</td><td>No.</td><td>↓↓(<1000)</td><td>↑↑</td></tr>
<tr><td>Type</td><td>Lymphocytes</td><td>PMNLs</td></tr>
<tr><td colspan="2">LDH</td><td>↓↓</td><td>↑↑</td></tr>
<tr><td colspan="2">Organisms</td><td>-ve</td><td>+ve</td></tr>
<tr><td colspan="2">AE</td><td>- Hepatic: LCF,, Portal HTN.
- Systemic:PEM,,↑↑Loss(NS,PLE).</td><td>- Infections(peritonitis).
- Mg: neuroblastoma.</td></tr>
</table></td></tr>
<tr><td>TTT</td><td>- AE.
- Monitor.
- Diet: ↓↓Na, ptn, salt.&↑ K.
- Diuretics.
- Transfusion: FFP, Albumin.
- Liver support.
- Paracentesis: diagnostic& therapeutic.</td></tr>
</table>

Cysts of liver& Biliary system

1- Choledochal cyst	- Cystic dilatation of CBD(Saccular/ fusiform). - Infancy: asymptomatic. - Cholestasis + abd.pain - TTT: Excision.
2- Caroli disease	- Multiple cystic dilatations of intra-hepatic ducts. - C/P: Acute cholangitis(fever, abd.pain,jaundice, pururitis). - TTT: Abs+ excision.
3- Cong. Hepatic fibrosis	- AR.(Diffuse peri-portal fibrosis). - C/P: HSM,,, Portal HTN+ Polycystic kidney(75%). - D: Biopsy,,,LFT. - TTT: LCF, RF, Portal HTN.
4- Polycystic kidney	- Infantile: AR,,,CRF,,,,Liver affection - Adult: AD
5- Hydatid disease	- Rare (Ecchynococcus granulosis,,,dogs).